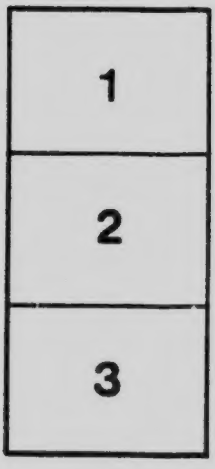

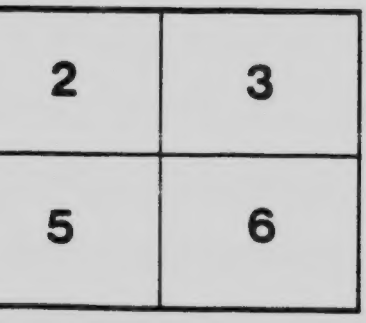

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By Frederic T. Lewis, Harvard Medical School, and Maude E. Abbott, McGill University.

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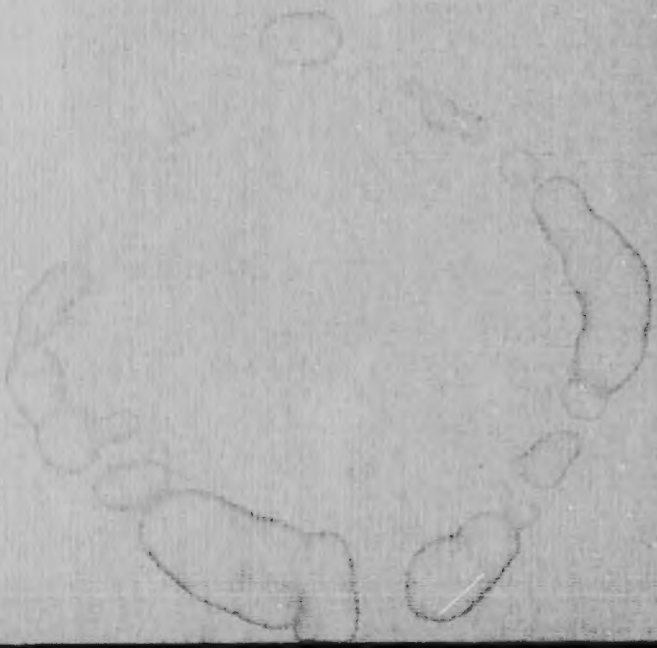

REVERSED TORSION OF THE VENTRICULAR BEND OF THE EMBRYONIC HEART IN THE EXPLANATION OF CERTAIN FORMS OF CARDIAC ANOMALY.*

FREDERIC T. LEWIS, HARVARD MEDICAL SCHOOL, AND MAUDE E. ABBOTT, McGILL UNIVERSITY.
(*Presented by Dr. Abbott*).

PRELIMINARY COMMUNICATION.

In the now quite well recognized anomaly known as transposition of the arterial trunks the aorta and pulmonary artery arise in more or less completely reversed relations; the aorta comes off anteriorly from the right ventricle and the pulmonary posteriorly from the left, the ventricles and auricles remaining as in the normal heart, that is, not sharing in the transposition. Such a state of things means of course a short circuiting of the circulation so that aerated blood is returned from the left ventricle to the lungs while venous blood is sent from the right ventricle back into the systemic circulation without aeration. It is naturally incompatible with life unless relieved by some compensating communication between the two circulations, such as is supplied by a patent ductus or foramen, or a defective inter-ventricular septum: even in these cases the condition leads at birth to the most extreme type of *morbus coeruleus* known.

The subject of transposition of the arterial trunks received its first elucidation from the great work of Rokitanski, who in his elaborate monograph on cardiac defects attributes it to a deviation of the spirally placed aortic septum. Developing in the primitive common arterial trunk at an early stage, the septum normally divides it into an anteriorly placed pulmonary artery, which is thrown into exclusive communication with the right ventricle by the union of the aortic and interventricular septa, and a posteriorly placed aorta, which is united with the left ventricle in the same manner. In the light of the more exact knowledge of today this theory does not seem to meet all the facts, although it probably has a place in their explanation. The subject has been further

* Presented before the American Association of Pathologists, and Exhibited at the American Section of the Museums Association, May, 1916.

studied by Dr. Arthur Keith and Dr. Jane Robertson[1], but the origin of this anomaly is still obscure.

During the course of a study of the recorded cases of congenital cardiac diseases made by one of us (M. E. A.) for a special purpose[2], our attention was attracted to a rare form of cardiac anomaly observed in a specimen in the Warren Anatomical Museum of the Harvard Medical School. In this specimen, as in six similar cases recorded in the literature (Young[3], Peacock[4], Rokitanski[5] (two cases), Theremin, obs. 43, and Marchand[6]), the great vessels arise in transposed relations, and a malposed muscular septum cuts off a small chamber which gives off the aorta and is situated at the upper and anterior aspect of the ventricular portion of the heart. The large remaining portion of the ventricle gives off a small thin-walled pulmonary artery, receives the mitral and tricuspid orifices, and communicates with the small chamber by a large diamond shaped opening. This heart is from a youth of twenty-one, who died in 1838 of pulmonary phthisis. The most striking thing in its appearance is the transposition of the great trunks: the large thick walled ventrally placed aorta emerges anteriorly from the small imperfectly divided-off portion of the ventricle, and crosses in front of the pulmonary artery, which arises behind, in the position of the normal aorta, from the main portion of the large common ventricle. The distal relations of the two trunks to the aortic arch, including the right and left pulmonary branches and ductus arteriosus were apparently normal, and the atria and atrio-ventricular orifices are normally arranged. That is to say a true transposition of the arterial trunks, without transposition of the heart as a whole, exists. In Fig. 1 this heart is shown at B beside a normal one for comparison (A). (Fig. 1, B).

A specimen in the Museum of McGill University represents an interesting variant of the above anomaly, in that the great trunks arise from the respective chambers in their normal instead of the transposed relations, thus proving that their transposition is not an essential part of the complicated anomaly presented by the Harvard heart and the like cases. This specimen, which in

[1] Journ. Path. and Bact. 1913, xviii, p. 191.

[2] Abbott, Osler and McCrae's System of Medicine, 2nd Edition, Vol. IV, 1915.

[3] Journ. Anat. and Physiol., 1907, xli, 190.

[4] Trans. Path. Soc., London, 1854, vi, 177.

[5] Vienna, 1875.

[6] Verh. d. XII Deut. path. Gesell, 1908, p. 174.

this particular is unique in the literature, was reported by the late Dr. Andrew Holmes[7], first Dean of the Medical Faculty, in 1823. It is figured diagrammatically in Fig. 2.

The Harvard specimen was brought to Dr. Frederic Lewis by the writer for embryological interpretation and formed the starting point of the present investigation. From the conditions seen in the Holmes heart it was clearly evident that the problem to be explained was a two-fold one, namely, (a) the peculiar form of triloculate heart common to all the cases in the series, and (b) the transposition of the great trunks complicating the present case. Dr. Lewis thought that the first of these two conditions probably represented an arrest of development at a stage shown in a model already made by himself of the interior of a 10 mm. embryo heart (Fig. I E.) and further suggested as an explanation of the transposition that in the embryo the cardiac tube had bent in the reverse direction to that which is normal, so that the aortic limb turned upward on the left side of the common ventricle instead of on the right, thus bringing the great vessels into an apparently reversed relation with the vessels of the arch. That such a reversed torsion of the cardiac tube can and does occur in life is shown in a plate in Remak's[8] atlas in which the heart of a chick embryo at a very early stage before pouching of the auricles has occurred, is seen abnormally bent to the left side, while a second heart at the same stage, with the normal bending of the tube to the right, is shown in the same plate for comparison.

In order to verify Dr. Lewis' suggestion of reversed torsion as an explanation of transposition, (which we found had already been made by Keith), he and the writer together undertook the following investigation. Normal embryonic hearts at the critical stages were selected and modeled as they occur in the embryo. From the same set of drawings second models were then made in which the ventricular portion of the heart was reversed, section by section. The two models of the heart of a 4.9 mm. embryo, kindly loaned to us by Dr. A. S. Begg, as also models of the exterior view of the heart of the 10 mm. human embryo have been completed in this way and are figured here (Fig. I, C-F). With others of the interior, which are still unfinished, they seem to demonstrate the correctness of the interpretation. The distal

[7] Edin. Med. Chir. Soc., 1824, republished by M. E. Abbott, Montreal Med. Jour., 1901.

[8] Remak, Untersuchungen über die Entw. der Wirbelthiere. Taf. iii, Figs. 28 and 29.

part of the aortic trunk, including the roots of the pulmonary and fourth aortic arches, remains undisturbed in all its relations, and the atria and atrio-ventricular orifices are also in essentially normal position. But the reversal of the primary torsion causes the aorta to be split off from the truncus arteriosus ventral to the pulmonary artery, around the front of which it swings to the left ventricle.

The interior view of the reversed model (Fig. I, F) is seen to resemble closely the appearance in the Harvard heart, in which an arrest of development at this stage is complicated by transposition, and on the other hand a comparison of the interior of the reconstruction from the same heart in its normal condition, i. e. unreversed, with the interior of the Holmes' heart, in which the great trunks arise in normal relations, shows strikingly similar features.

The application of this theory of the reversal of the cardiac tube as an explanation of transposition of the arterial trunks in the comparatively large group of cases in which this occurs, uncomplicated by the anomaly seen in the Harvard heart, is less obvious. For, if the reversal be complete, and development proceeds on this basis, one must conclude that, the relationship of all the parts concerned being entirely reversed, no pathological results would ensue, that is to say, the aorta, although placed anteriorly, would continue to drain the left ventricle and the pulmonary artery, although placed posteriorly, would continue to drain the right. The displacement of the orifices relatively to the pulmonary and aortic trunks and to the atrio-ventricular orifices would take effect only when the reversed torsion was incomplete, so that from juxta-position the aorta would be thrown into relation with the mitral orifice and the pulmonary with the tricuspid. That such an incomplete torsion is the underlying cause in the cases of true transposition is borne out by the examination of two cases, kindly loaned to us for purposes of study by Professor W. G. MacCallum in New York, and figured by him in the New York Pathological Society Transactions for 1915. In both these specimens the aorta arises anteriorly and slightly to the right from the ill-developed conus of the right ventricle and passes upward to the arch, while the pulmonary artery arises from the left ventricle, to the left and only slightly posteriorly to the aorta, being almost on a parallel plane with it. This is not a complete reversal of the two trunks in the sense that although they arise from reversed ventricles they do not take up the relative position normally occupied by each other. An exactly similar external appear-

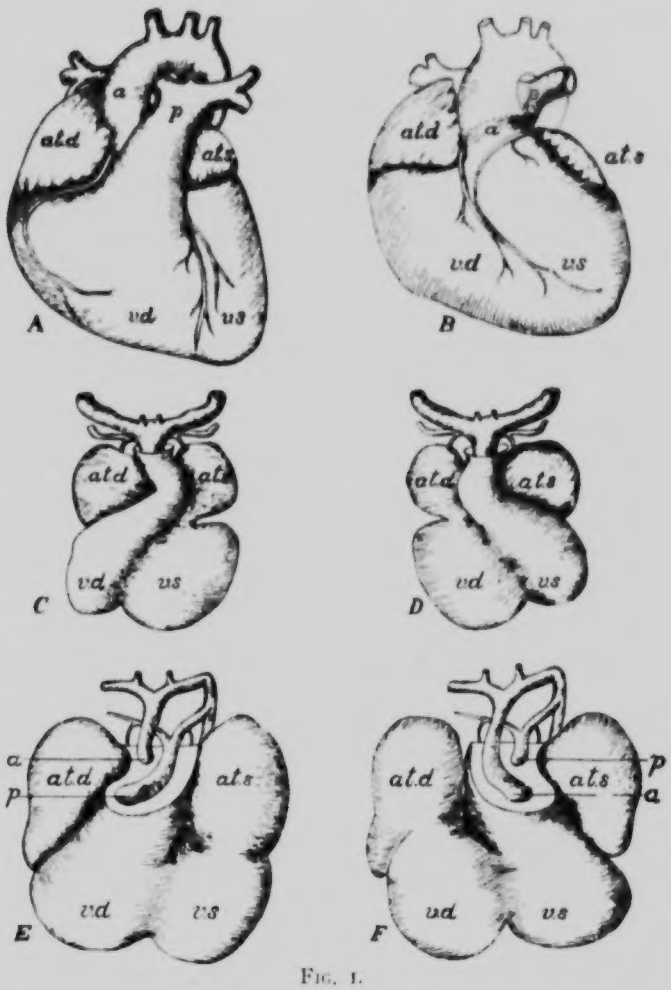

FIG. 1.

A, ventral view of a normal adult human heart. *B*, corresponding view of an adult human heart showing reversed torsion. *C*, model of the heart of a 4.9 human embryo, which in *D* has been manipulated so as to present reversed torsion. *E*, model of the normal heart of a 10 mm. human embryo, the torsion of which has been reversed in *F*.

a, aorta. *at. d*, right atrium (or auricle). *at. s*, left atrium (or auricle). *p*, pulmonary artery. *v. d*, right ventricle. *v. s*, left ventricle.

Reprinted from the Anatomical Record. Lewis and Abbott, 1915. Vol. 9.

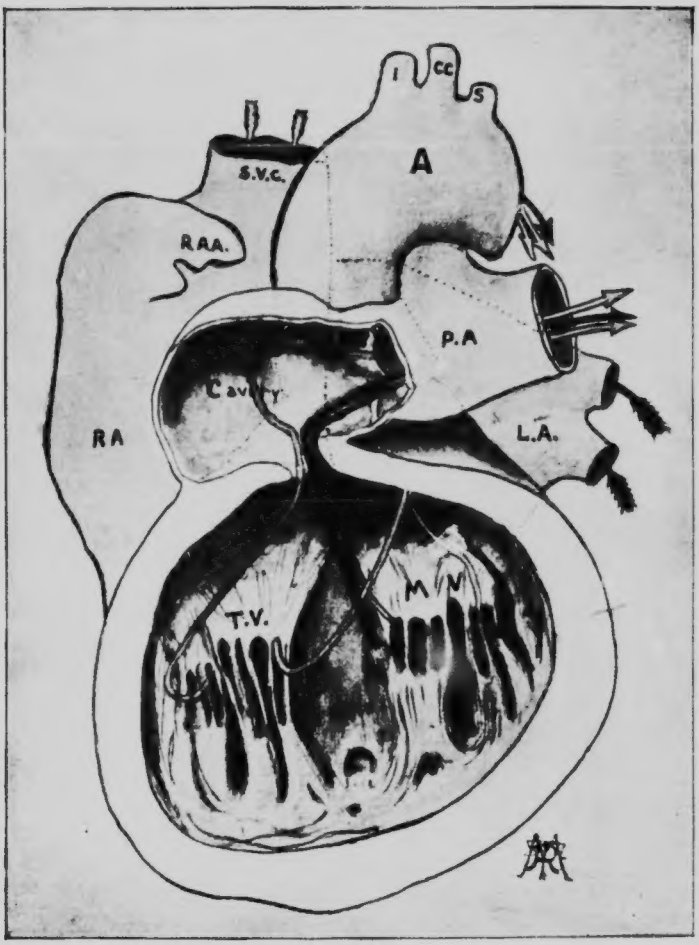

FIG. 2. Cor Biatriatum Triloculare with malposed interventricular septum, cutting off a small chamber which gives off the pulmonary artery from a large common ventricle which gives off the aorta.

Note that the great vessels are here in their normal relations, i. e., are not transposed. The pale line shows venous, the dark line arterial blood.

Diagrammatic sketch by Prof. R. Tait Mackenzie, from a specimen in the Medical Museum of McGill University, presented by Dr. Andrew Holmes, first Dean of the faculty in 1824.

ance is produced in the embryonic hearts reconstructed, if instead of complete reversing the ventricular portion of the plates, a partial reversal is carried out; such a partial reversal interiorly should have the effect of throwing the aorta into communication with the right ventricle and the pulmonary artery with the left. Two such anomalous models have been made by us, illustrative of an incomplete torsion both of the normal and of the reversed cardiac loop. In both of these cases it may be supposed that the aorta would be thrown into the right ventricle and the puimonary to the left, although the external relations of the two vessels would differ in each case.

The investigation upon this interesting subject, especially in relation to the interior conditions arising as a result of such deformities is being completed and will be published at a later date.